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Benign vascular malformation of the diaphragm☆

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ABSTRACT

Lesions of the diaphragm are rare and often ominous neoplasms with a broad range of presenting symptoms. While benign diaphragmatic masses have been described, they comprise the minority of these lesions and imaging is ineffective at differentiating between the two. We describe an unusual case of a 15 year-old girl presenting with a benign vascular malformation of the diaphragm.

1. Case report

A 15-year-old female presented with 3 days of right upper quadrant pain and associated nausea. Her pain began suddenly after eating and was exacerbated in the supine position and with deep inspiration. She denied a history of similar pain, cough, or trauma to the region. She was afebrile and constitutionally well.

Abdominal exam revealed tenderness, which was most intense at her right costal margin and radiated to her shoulder. Biochemical analysis and abdominal ultrasound were within normal limits with the exception of isolated splenomegaly. An MRI and CT scan demonstrated a 2.8 × 1.9 cm mass posterior to the 5th/6th intercostal space with a small pleural effusion (Figs. 1 and 2). Given the concern for malignant process, operative intervention was pursued.

A laparotomy was performed through a right subcostal incision. Intraoperative exploration revealed a 3 cm firm, bulging irregularity on the peritoneal surface of the diaphragm. Dissection of the mass resulted in violation of the pleura and pleural fluid was sent for cytology. The mass was situated on the pleural surface of the diaphragm (Fig. 3). Gross total resection was performed and frozen section was consistent with a benign vascular malformation (Figs. 4 and 5). A chest tube was placed and her diaphragm was closed primarily.

2. Discussion

Diaphragmatic masses are rare, represented in the literature by case reports and small series. The differential diagnosis include congenital bronchopulmonary malformations such as pulmonary sequestration [1] and bronchogenic cysts [2,3], vascular malformations such as hemangiomas [4] as well as malignancies of predominantly mesenchymal origin (inflammatory myofibroblastoma [5], solitary fibrous tumors of the pleura [6], rhabdomyosarcoma [7]). In the largest review of the literature, Cada et al. identified fewer than 200 cases over approximately 140 years, of which, 78% were malignant [8].

Of particular challenge in the diagnostic evaluation of diaphragmatic masses is the limitation of imaging to help determine etiology as well as location surface specifics [9] (peritoneal versus pleural). Traditional imaging modalities such as ultrasound, CT and MRI have all been employed, either in isolation or combination. However, none of these have been able to reliably determine location with any significant degree of sensitivity of specificity [8]. In the case presented, the mass was determined to be of diaphragmatic origin on both MRI and CT and was thought to be located on the peritoneal surface of the diaphragm. An abdominal approach was chosen as it was felt to have more optimal visualization and allow an opportunity to survey the peritoneal and

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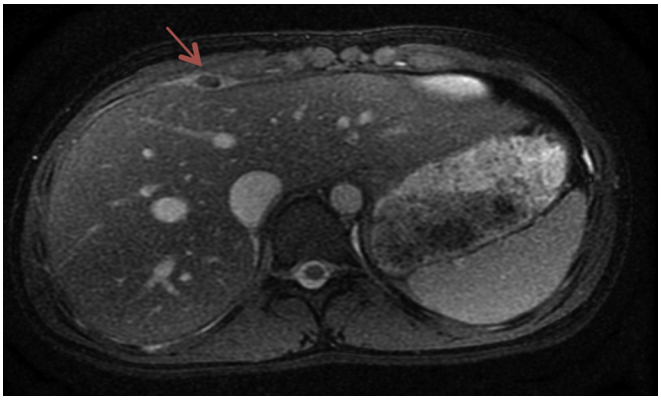

Fig. 1. Axial MRI image with small focus of decreased T1 and T2 signal between anterior aspect of liver and right chest wall (arrow).

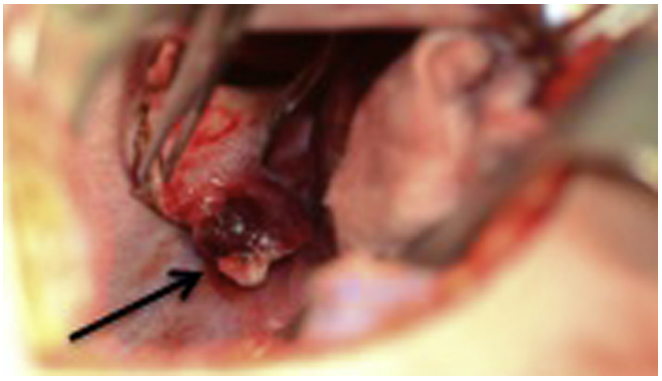

Fig. 3. Pedunculated mass on the peritoneal surface of the diaphragm (arrow).

hepatic capsular surfaces. A minimally invasive thoracic approach would have been equally feasible in this situation were the lesion felt to be on the pleural surface of the diaphragm.

With respect to intraoperative management, one must adhere to the basic oncologic tenets of en block resection with appropriate margins. Frozen section analysis is useful in determining the extent of resection required. There remain several options for diaphragmatic reconstruction after resection, including primary repair and several prosthetic mesh options, such as a GoreTex patch. Primary closure is optimal if margins are clear or, in the case of a benign lesions, as long as closure under tension is avoided.

3. Conclusion

The diagnosis of a primary diaphragmatic mass is inherently challenging due to nonspecific clinical symptoms and limitations of imaging. This may lead to unnecessary work-up and ultimately delay definitive treatment. The importance of pursuing an accurate diagnosis must be emphasized given the degree of concern for malignant tumor. Diaphragmatic lesions, both benign and malignant, should be considered in the differential diagnosis of pediatric patients presenting with either thoracic or abdominal complaints

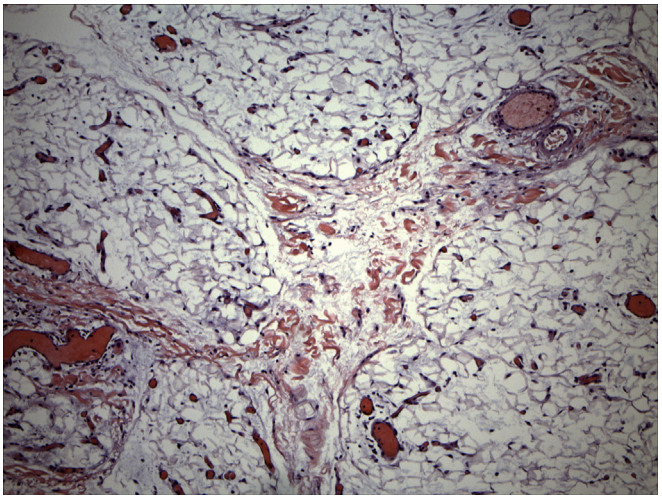

Fig. 4. Microscopic examination revealed an ill-defined lesion predominantly composed of adipose tissue with portions of a reactive lymph node. Numerous benign capillaries were seen admixed with the adipose tissue (10×, H&E).

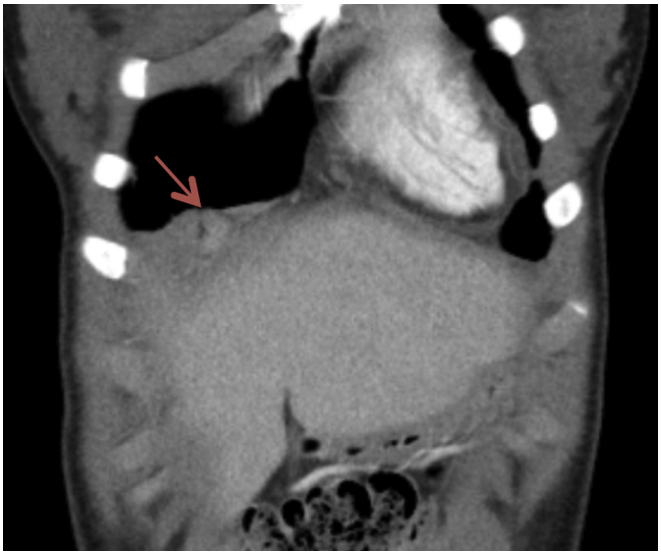

Fig. 2. Coronal CT with contrast demonstrating a focal abnormality with central hypoattenuation (arrow) and a small right pleural effusion.

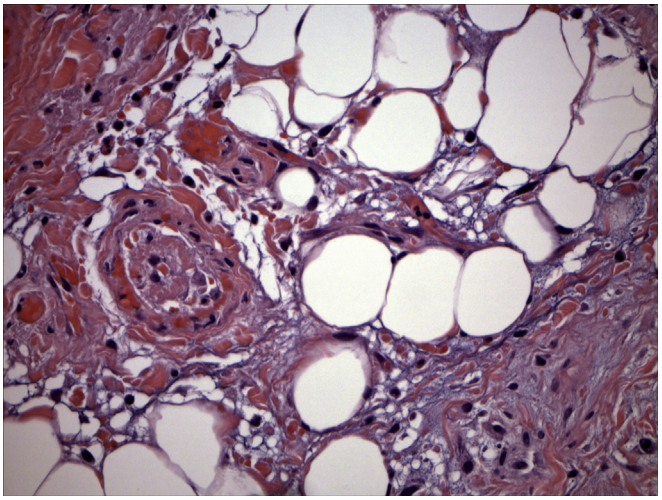

Fig. 5. Diagnosis of benign vascular lesion was favored due to the presence of increased vascularity, rare capillaries with fibrin thrombi and associated hemorrhagic infarction (20×, H&E) with angiolipoma considered in the differential diagnosis.

and cross sectional imaging may be helpful when such a lesion is suspected. Tissue diagnosis should be pursued with frozen section to guide intraoperative management.

Conflicts of interests and source of funding conclusion

None.

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